DESMOID TUMORS OF THE ABDOMINAL WALL.

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THE term "desmoid" was first used by Johannes Mueller [1] with application to certain tumors of connective tissue origin, and as the name implies, of "tendon-like" consistency, arising from the abdominal wall. These tumors are so unusual, and present such interest and possible difficulty in diagnosis, that the recent occurrence of a case in the clinic of Dr. Watts, at the University of Virginia, suggested the advisability of placing the case on record, with a brief résumé of the literature of the subject.

It is well, first of all, to determine what shall, and what shall not be included under the term desmoid. There have been some who would so classify the fibro-myomata of the round ligaments arising in the canal of Nuck, but essentially analogous to the ordinary uterine myomata. Similarly, the tumors to which attention was called by Nélaton,[2] arising from the bony pelvis and invading the abdominal wall, have been considered desmoids; but the weight of opinion is decidedly toward excluding these classes of tumors from the category of true desmoids. The conception of Pfeiffer,[3] which is also that authorized in v. Bergmann's "Handbuch der Praktischen Chirurgie," would restrict the use of the word "desmoid" to fibromata or fibro-sarcomata arising from the musculo-aponeurotic structures—muscles, muscle-sheaths, aponeuroses, lineæ transversæ, etc.—of the abdominal wall itself, thus excluding tumors originating in the bony pelvis or the round ligaments, as well as those springing from the skin or subcutaneous tissues.

In reviewing the literature, one is struck by the fact that the great majority of the articles on the subject are reports of single, or at most, three or four cases. There are, however,

two prominent exceptions to this general rule, in the monographs of Ledderhose[4] and Pfeiffer.[3] The first of these writers has collected 100 cases from various sources; the second reports 40 cases of his own, to which he adds 360 more cases collected from the literature, including the 100 cases of Ledderhose already mentioned, thus making a total of 400 cases in all. So thoroughly has the last author worked over the literature up to the date of his publication, that we feel it unnecessary to do more than refer the reader to this article with its voluminous bibliography, for work done before 1904. Since that date, we have been able to collect the following cases, which we report here in brief abstract.

CULLEN[6]: Mrs. N. M., 30 years old. No note of pregnancy. Tumor in left hypochondrium. Tumor lobulated and freely movable. At operation it was found to be attached to the sheath of the abdominal muscles, and in removing it, some of the muscle was taken out with the tumor. Pathological report: pure fibroma.

SCHWARZSCHILD[6]: Case I. Woman 28 years old. Tumor present in right side of abdomen for 2 years; growth very slow. For past half year, following directly upon labor, growth has been more rapid. Tumor is now the size of a child's head, hard and nodular, and attached to abdominal wall. Diagnosis: fibroma. At operation, the tumor was found to be closely adherent to peritoneum and a large defect was left by its removal, which was closed by a plastic operation. After one year, no recurrence, no hernia. Pathological report not made.

Case II. Woman, 30 years old, mother of two children. In the right lower quadrant of abdomen is a tumor, the size of a hen's egg, which is attached to abdominal wall, but not adherent to the skin. At operation the tumor was found to spring from the posterior surface of rectus, but was not adherent to peritoneum, which was not opened. Diagnosis: desmoid.

EITEL[7]: Mrs. J., aged 26 years. Tumor above and to left of symphisis pubis, of 10 months' duration and slow growth. Slight pain. Tumor size of fist, hard, encapsulated, deeply embedded in abdominal wall. At operation, tumor was found just beneath external oblique muscle, involving all the structures down to the peritoneum, to which it was adherent. Apparently tumor originated from fascia transversalis. Good closure without plastic. Patient now well. Pathological report: very cellular fibroma.

GROSS AND SENCERT[6]: Woman 73 years old. Tumor of 10 years' standing, arising to left of middle line and occupying whole left half of abdomen. Tumor very large, weighing 6 kg., and extensively ulcerated. Patient in condition of septicæmia. Tumor found at operation to be

attached by pedicle to the anterior sheath of rectus, and originating at the position of one of the lineæ transversæ. Pathological report: partly gangrenous lipo-fibroma.

ECCLES[9] and BEDWELL[10] report several cases of fibro-sarcomata of the abdominal wall, and another case is reported by TAPIE and DAUNIC[11] but the writer was unfortunately unable to gain access to these articles.

Besides these cases, five others have been collected from the surgical service of the Johns Hopkins Hospital, for the permission to use which we wish to thank Dr. Wm. S. Halsted and Dr. Jos. C. Bloodgood of that institution. These cases were grouped under the caption "tumors of the abdominal wall," and were collected from a general surgical service now exceeding 21,000 cases of all kinds. The abstracts follow.

CASE I.—A man, aged 27, with tumor just below umbilicus in mid-line. Duration 6 years. During the past 5 months, more rapid growth has been observed. Tumor is hard, smooth, just under skin, freely movable. Tumor easily enucleated. No statement as to attachment of tumor to deep structures. Microscopically, the tumor is composed largely of spindle cells.

CASE II.—Woman, aged 56. Has borne a child. Shortly after labor, a painless, slow-growing tumor appeared just to left of umbilicus. Tumor was incompletely removed, the wound became infected, and healed with a large scar. For the past 20 years there has been a gradual recurrence in this scar, the tumor involving the skin. For five years, the tumor has been ulcerated, which patient attributes to trauma. Lately two small secondary nodules have appeared in skin to right and left of original growth. Tumors are hard, freely movable on deep structures, but firmly fixed to the skin. Tumor easily removed, and no note made of any attachment to deep structures. Pathological report: spindle-celled tumor.

CASE III.—Woman, aged 23 years, with tumor of 6 months duration, onset following pregnancy. Tumor to the left and below umbilicus, is palpable but not visible. Tenderness prevents accurate palpation, or definite outlining of mass. Fixed to deeper structures. At operation, tumor is found to be attached to posterior surface of the sheath of rectus.

CASE IV.—Woman, aged 35 years, with rapidly growing tumor of the right lower quadrant of abdomen, following pregnancy. No pain. Tumor is hard, nodular, with skin freely movable, and itself movable on deep parts. At operation, tumor is described as subcutaneous, but note states that part of rectus had to be removed with it, because of its firm adherence to the anterior sheath of that muscle.

CASE V.—Colored woman, aged 18. Tumor appeared during pregnancy, to left of and just below umbilicus. Only a few months duration.

Tumor is hard, somewhat nodular, and seems to be beneath rectus sheath. At operation, tumor is found under the anterior sheath of rectus, and infiltrating the muscle. Pathological report: spindle-celled sarcoma.

At this point we would enter a brief commentary upon these five cases. Case I may or may not have been a desmoid. Neither the clinical findings, nor the operation note, make clear its point of origin, and it may perfectly well have been an ordinary subcutaneous fibroid nodule. It must bear the verdict "not proven." Case II similarly is not above suspicion. Whatever the nature of the primary growth, which was incompletely removed long before the patient sought the hospital, the recurrence in the scar certainly bears much closer resemblance to a keloid or sarcoma of the skin than to a true desmoid. As to the last three cases, we can undoubtedly regard them as cases for inclusion in the class of tumors under consideration. We would like to draw attention to Case V, as far as we know a unique example of true desmoid in the negro, contrasting strongly with the frequent occurrence in this race of the keloid, a tumor certainly very closely related to the desmoid.

Finally, we have to add to the cases here compiled, one which has recently occurred in our own clinic, and which led primarily to this review of the subject.

MRS. A. G., widow, aged 23 years, entered the hospital complaining of an abdominal tumor. Her family history is of no importance. The striking fact in her past history is the instrumental delivery of a still-born child, said to have been unusually large. This was the only pregnancy. The present illness began seven months ago, when patient noticed a lump in the left half of the abdomen. There has been no apparent growth since discovery. No pain or other subjective symptoms of any kind. On examination, the patient's general condition is excellent. No organic lesions. Patient quite stout. Abdomen is full, soft, with thick fatty walls. Nowhere tender. About 2 cm. to left of the umbilicus, there is an ill-defined, deep-lying, ovoid mass, measuring about 8 cm x 5 cm., with its long axis parallel to the fibres of the rectus. It is firm, smooth, and the skin over it is freely

movable. The mass itself moves with the abdominal wall. No apparent connection with the pelvis, or the internal genitals. Vaginal examination throws no light on the nature of the tumor. No other masses anywhere.

A definite diagnosis was not made. The very thick abdominal walls gave the impression that the tumor was intra-abdominal,—in fact it was partly so,—and the possibility of an omental or mesenteric growth, perhaps with adhesion to the anterior abdominal wall was considered.

At operation, the tumor was found to be a desmoid, arising from the posterior sheath of the left rectus, involving the entire thickness of the muscle, and the median two-thirds of its width. The greater mass of the tumor projected backward into the abdominal cavity, to such an extent that the bulk of the tumor was intra-abdominal, and was firmly adherent to the peritoneum. In excising the growth, a piece of peritoneum 4 x 5 cm. was removed with it. The abdominal contents were normal. Closure was effected in layers, and a strong repair made without the necessity of any plastic measures. Pathological report: very cellular fibroma.

In reviewing the cases above reported, with those collected by other authors, certain salient features have been observed in regard to desmoids, which are commented upon by all the writers on the subject. These characteristics we will proceed to outline, using Pfeiffer's work freely for statistics.

Pathology.—These tumors are of connective tissue origin and spring from the musculo-aponeurotic structures of the abdominal wall. In the gross they are hard tumors, occasionally with areas of softening from cystic degeneration; smooth or slightly nodular in outline; cut with a crisp grating, and on the cut surface present a dense fibrous structure. Microscopically the majority of specimens present the typical picture of a more or less cellular fibroma. In a certain number of cases, careful sectioning shows areas of sarcomatous change, and a few are pure sarcomata. Other variations occasionally met with are tumors presenting areas of myxomatous or

hemorrhagic degeneration. These facts have led to the use of various compound names, *i.e.*, fibro-myxo-sarcoma, etc., but the best authorities sustain the practice of Sänger who groups all these tumors under the one term desmoid. The former generally accepted belief in the rarity of malignant tumors of this class has been considerably modified by the statistics of Pfeiffer [8] in whose tabulation 10.6 per cent. of the cases in women and 24.4 per cent. of those in men were sarcomata. This large proportion of malignant cases in the male is worthy of note, as is also the curious fact, that the clinical and microscopic evidences of malignancy show less harmony in this class of tumors than perhaps any other. Tumors which show rapid growth, invasion of neighboring parts, and pain, not infrequently are pure fibromata; whereas, on the other hand, clinically benign, quiescent growths may present typical fields of sarcoma under the microscope.

Incidence.—The rarity of these tumors may be appreciated from the statistics of Guerlt, obtained from the Vienna hospitals, .13 per cent. of desmoids in 16,637 tumor cases. Perhaps the most striking peculiarity of the desmoid is the preponderance of its occurrence in women, and particularly parous women. Nor infrequently the tumor is first discovered during pregnancy or the puerperium. To have recourse again to Pfeiffer's figures, he shows that 87.1 per cent. of his cases were in women, and that 94.3 per cent. of the women had borne children. The tumors may occur at any age from 1½ to 81 years. In fact, a rare case or two, considered congenital, is on record. The period of life of greatest liability is from 25 to 35 years in women and 35 to 50 in men.

Location.—The most frequent position of desmoids is in the right lower quadrant of the abdomen. The anatomical structure from which they most frequently arise is the rectus abdominis muscle, or its attachments, sheath, lineæ transversæ, etc. Next in order of frequency come the external oblique muscles, the fascia transversalis, and the lineæ alba. A characteristic of the tumors, which are usually ovoid, is that their long axes are nearly always parallel with the direction of the

fibres of the muscle in which they are growing, so that desmoids in the middle of the abdomen lie longitudinally, whereas those in the flanks are transverse. Nélaton [2] believed that these tumors frequently originated from the bony pelvis, and that most of them were connected with it by a fibrous pedicle, but Guyon [12] has shown that such a connection either does not exist at all, or is simply a band of fascia under tension. Desmoid tumors are solitary; at least, multiplicity has never been proved.

Etiology.—The peculiarities of desmoids have led to much speculation and discussion as to whether they may not have some special causation aside from those factors that may lead to tumor growth elsewhere. The fact to which attention was drawn above, namely, that pregnancy seems to bear some relation to the incidence of these neoplasms, and the further fact, that many cases occurring in men or nulliparous women, give a history of preceding trauma, has furnished ground for much speculation. Herzog [13] and others support the theory that during pregnancy or parturition there is a rupture of the structures of the abdominal wall that leads to a fibrous scar or a hematoma. This scar or organizing hematoma is conceived to be the starting point of a desmoid, in much the same way as a skin scar is the starting point of a keloid. Others have supposed that the stretching of the muscles of the belly-wall during pregnancy plays an important part in the process, and explain the striking absence of desmoids in cases of distention from ascites, ovarian cysts, etc., on the ground that in these conditions the blood-supply to the abdomen is impaired, and the general nutritive resources of all the tissues is low, whereas in pregnancy, just the reverse is the case. Certain experimental work on pregnant and ascitic animals lends color to this reasoning. But while pregnancy may present favorable conditions for desmoid formation, the not infrequent occurrence of such tumors in cases with no history of either pregnancy or trauma, makes it probable that there is some other more important factor in the etiology, and the general belief is that the cause of desmoids will be explained only

when the mystery, that as yet enshrouds neoplasms in general, is finally solved.

Clinical Course.—In the majority of cases, the patient accidentally discovers the existence of the mass. Pain or subjective symptoms of any kind are unusual; if the growth be quite large, there may be dragging, aching sensations, or the tumor if large and properly situated may give rise to visceral disturbances from pressure, the bladder being the organ most frequently involved. The tumors, when first discovered, are usually from about the size of a hen's egg to that of a clenched fist. In most cases growth is slow, possibly imperceptible. Calcification may put a stop to the progress of the tumor. In some cases, however, growth may be quite rapid, the tumor reaching the size of a child's or even an adult's head in a few months. Recession and spontaneous disappearance of a true desmoid has never been observed. The larger tumors, particularly if projecting anteriorly, are liable to traumatism or friction from the clothing, and as the skin over them is tense and thin, with dilated veins, conditions favorable for ulceration exist. When this occurs, a portal of entry for infection is of course opened, and death from this cause is a well recognized termination of large desmoids.

The lymph glands usually are not involved unless the tumor is of a most malignant type. As has been stated above, clinical indications that suggest malignancy, such as recurrence after apparently complete removal, or invasion of surrounding parts, may occur in tumors microscopically benign. It should be noted in passing that such "invasion" is really rather a pushing aside of the neighboring tissues, since although these tumors seldom have a definite capsule, they are well circumscribed, and neither clinically nor microscopically tend to diffuse permeation of the tissues, except in cases of pure sarcoma. Let me again, however, call attention to the figures given above regarding the frequency of sarcoma, particularly in men, and emphasize the mistake made in the past of attributing so little possibility of malignancy to desmoids.

Diagnosis.—One would think there would be little diffi-

culty in recognizing these growths, and in many cases this is true; but where the patient has thick abdominal walls, and the tumor is deeply situated, it is by no means easy. A tumor springing from the anterior sheath of the rectus is usually easy to diagnose. Such a tumor, which is freely movable when the abdomen is relaxed, disappears or becomes fixed when the muscles are made tense by straining, or raising the head from the pillow without the help of the arms. From cases lying within the muscle, suppuration, hematoma, and cysts of various kinds have to be differentiated. The tenderness and other signs of inflammation usually render the first of these problems easy. Cysts may be diagnosed by aspiration, particularly if this possibility is suggested by fluctuation, which is not, however, always present in cysts. In deep lying hematomata with firm tense capsules the findings may be most confusing, but the fact that a hematoma either is absorbed or suppurates, whereas a tumor grows, will distinguish the two lesions if one has opportunity to observe the case for a time, or can secure a trustworthy history.

Lastly, and most difficult of all to diagnose, are those cases in which the tumors project posteriorly into the abdominal cavity. Here one has to consider the possibility of the tumor being of visceral origin, and if the walls be thick, palpation is most unsatisfactory. Tumors of the liver may be ruled out by the descent of that organ with inspiration. The spleen usually has a characteristic edge, but certain cases may be most confusing. Kidney tumors can usually be ruled out by the change in percussion and palpation following inflation of the bowel. Intestinal growths give rise to symptoms which are entirely unlike the desmoid picture, and are besides usually mobile and take up different positions. Finally, the rare tumors, especially sarcomata, of the omentum or mesentery, may present great difficulty, and indeed, if they are adherent to the anterior abdominal wall, the distinction may be impossible to make.

Treatment.—The question of what to do for these growths, may be answered in two words: *operate early.* The

not remote possibility of malignant degeneration in any tumor of this class is sufficient reason for such advice, but aside from this the direction of growth of many desmoids furnishes another strong reason. All of the cases which spring from the posterior wall and grow backward naturally become closely applied and adherent to the peritoneum. Furthermore, in their extension laterally they either cause pressure atrophy of the muscles, or push them aside. The longer such a condition lasts, and the further it extends, the larger defect is made both in the muscular and peritoneal layers, by the complete removal of the tumor. We have not space to describe the ingenious plastic methods employed in the closure of wounds by some of the surgeons who have removed large tumors; but we feel sure that one who has been forced to such resorts will afterward be a vigorous advocate of early operation. The chief factors that prevent perfect results are the occurrence of post-operative herniæ and recurrences of the tumor. That we may emphasize the gravity of the condition, we present the following statistics collected since the introduction of antisepsis.

Mortality in laparotomy cases.....3.5 per cent.
Mortality without laparotomy.....1.05 per cent.

Recurrences in men .....68.1 per cent.
Recurrences in women .....90.0 per cent.

Final cure, surviving 1st and possibly 2nd and 3rd operations:
Men .....50 per cent.
Women .....21.2 per cent.

In conclusion, I wish to express my gratitude and appreciation to my chief, Dr. Watts, for his stimulating support in the preparation of this paper.

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